SEQUENCE LISTING <110> CANFIELD, WILLIAM M <120> METHODS FOR PRODUCING HIGHLY PHOSPHORYLATED LYSOSOMAL HYDROLASES <130> 210119US0CONT <150> 60/153,831 <151> 1999-09-14 <150> US 09/635,872 <151> 2000-08-10 <160> 52 <170> PatentIn version 3.1 <210> 1 <211> 928 <212> PRT <213> Homo sapiens <400> 1

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Trp ∰al Asn Gly Thr Asp Leu Glu Leu Lys Glu Leu Gln Gln Val

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Asp Ala His Ser Gly Leu Leu Lys Gly Asn Ser Arg Gln Thr Val Trp 195 200 205

Arg Gly Tyr Leu Thr Thr Asp Lys Glu Val Pro Gly Leu Val Leu Met 210 215 220

Gln Asp Leu Ala Phe Leu Ser Gly Phe Pro Pro Thr Phe Lys Glu Thr

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Leu Leu Gln Leu Tyr Ser Glu Ala Ser Val Ala Leu Leu Lys Leu Asn 260 265 270

Asn Pro Lys Asp Phe Gln Glu Leu Asn Lys Gln Thr Lys Lys Asn Met 275 280 285

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Pro Asp Asp Phe Tyr Ser His Ser Lys Gly Gln Lys Val Tyr Leu Thr 420 425 430

Trp Pro Val Pro Asn Cys Ala Glu Gly Cys Pro Gly Ser Trp Ile Lys 435 440 445

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Gln Val His Lys Ser Ile Leu Pro Asn Ser Leu Gly Val Ser Glu Arg 770 780

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Arg Val Glu Thr His Thr Gln Lys Thr Ile Gly Gly Asn Val Thr Lys 820 825 830

Glu Tys Pro Pro Ser Leu Ile Val Pro Leu Glu Ser Gln Met Thr Lys

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Leu lin His Tyr Thr Asp Ser Tyr Leu Gly Phe Leu Pro Trp Glu Lys 885 890 895

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Asn Ile Asp His Asn His Lys Asp Ala Gln Thr Val Lys Ala Val Leu 225 230 235 240

Arg Asp Phe Tyr Glu Ser Met Phe Pro Ile Pro Ser Gln Phe Glu Leu 245 250 255

Pro Arg Glu Tyr Arg Asn Arg Phe Leu His Met His Glu Leu Gln Glu 260 265 270

Trp Arg Ala Tyr Arg Asp Lys Leu Lys Phe Trp Thr His Cys Val Leu 275 280 285

Ala Thr Leu Ile Met Phe Thr Ile Phe Ser Phe Phe Ala Glu Gln Leu 290 295 300

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Phe Arg Leu Ser Gly Lys Cys Phe Ser Leu Val Glu Ser Thr Tyr Lys 65 70 75 80

Tyr Clu Phe Cys Pro Phe His Asn Val Thr Gln His Glu Gln Thr Phe 85 90 95

Argi Trp Asn Ala Tyr Ser Gly Ile Leu Gly Ile Trp His Glu Trp Glu
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Ile Ala Asn Asn Thr Phe Thr Gly Met Trp Met Arg Asp Gly Asp Ala 115 120 125

Cys Arg Ser Arg Ser Arg Gln Ser Lys Val Glu Leu Ala Cys Gly Lys 130 135 140

Ser Asn Arg Leu Ala His Val Ser Glu Pro Ser Thr Cys Val Tyr Ala 145 150 155 160

Leu Thr Phe Glu Thr Pro Leu Val Cys His Pro His Ala Leu Leu Val 165 170 175

Tyr Pro Thr Leu Pro Glu Ala Leu Gln Arg Gln Trp Asp Gln Val Glu 180 185 190

Gln Asp Leu Ala Asp Glu Leu Ile Thr Pro Gln Gly His Glu Lys Leu Leu Arg Thr Leu Phe Glu Asp Ala Gly Tyr Leu Lys Thr Pro Glu Glu 215 Asn Glu Pro Thr Gln Leu Glu Gly Gly Pro Asp Ser Leu Gly Phe Glu 230 235 Thr Leu Glu Asn Cys Arg Lys Ala His Lys Glu Leu Ser Lys Glu Ile 245 Lys Arg Leu Lys Gly Leu Leu Thr Gln His Gly Ile Pro Tyr Thr Arg Pro Thr Glu Thr Ser Asn Leu Glu His Leu Gly His Glu Thr Pro Arg O Ala Bys Ser Pro Glu Gln Leu Arg Gly Asp Pro Gly Leu Arg Gly Ser 295 300 <u>29</u>0 Uī Leu 305 ΠIJ 4 <211> 5597 <212> DNA <213> Homo sapiens <400> 4 <210>-60 cggagccagag cgggcgtccg tcgccggagc tgcaatgagc ggcgcccgga ggctgtgacc 120 tgcg by ggc ggcccgaccg gggcccctga atggcggctc gctgaggcgg cggcggcggc ggcggttcag gctcctcggg gcgtggcgtg gcggtgaagg ggtgatgctg ttcaagctcc 180 tgcagagaca aacctatace tgcctgtccc acaggtatgg gctctacgtg tgcttcttgg 240 gcgtcgttgt caccatcgtc tccgccttcc agttcggaga ggtggttctg gaatggagcc 300 gagatcaata ccatgttttg tttgattcct atagagacaa tattgctgga aagtcctttc 360 agaatcggct ttgtctgccc atgccgattg acgttgttta cacctgggtg aatggcacag 420 atcttgaact actgaaggaa ctacagcagg tcagagaaca gatggaggag gagcagaaag 480 caatgagaga aatcettggg aaaaacacaa cggaacctac taagaagagt gagaagcagt 540 600 tagaqtqttt qctaacacac tqcattaaqq tqccaatqct tqtactqqac ccaqcctqc 660 cagccaacat caccctgaag gacgtgccat ctctttatcc ttctttcat tctgccagtg acattttcaa tgttgcaaaa ccaaaaaacc cttctaccaa tgtctcagtt gttgtttttg 720 acagtactaa ggatgttgaa gatgcccact ctggactgct taaaggaaat agcagacaga 780 840 cagtatggag ggggtacttg acaacagata aagaagtccc tggattagtg ctaatgcaag

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Pro Pro Pro Pro Ala Thr Pro Gly Ala Gly Gly Leu Ala Val Arg Thr 65 70 75 80

Phe Val Ser His Phe Arg Asp Arg Ala Val Ala Gly His Leu Thr Arg 85 90 95

Ala Val Glu Pro Leu Arg Thr Phe Ser Val Leu Glu Pro Gly Gly Pro 100 105 110

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Ile Asp Arg Ile Val Met Gl<br/>n Glu Leu Gl<br/>n Asp Met Phe Pro Glu Glu 35 40 45

Phe Asp Lys Thr Ser Phe His Lys Val Arg His Ser Glu Asp Met Gln

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Phe Ala Phe Ser Tyr Phe Tyr Tyr Leu Met Ser Ala Val Gln Pro Leu 65 70 75 80

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Asn Ile Ser Gln Val Phe His Glu Val Asp Thr Asp Gln Ser Gly Val 85 90 95

Leu Ser Asp Arg Glu Ile Arg Thr Leu Ala Thr Arg Ile His Asp Leu 100 105 110

Pro Leu Ser Leu Gln Asp Leu Thr Gly Leu Glu His Met Leu Ile Asn 115 120 125

Cys Ser Lys Met Leu Pro Ala Asn Ile Thr Gln Leu Asn Asn Ile Pro 130 135 140

Pro thr Gln Glu Ala Tyr Tyr Asp Pro Asn Leu Pro Pro Val Thr Lys
145 m 150 155 160

Ser Teu Val Thr Asn Cys Lys Pro Val Thr Asp Lys Ile His Lys Ala

165
170
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Tyr Lys Asp Lys Asn Lys Tyr Arg Phe Glu Ile Met Gly Glu Glu Glu 180 185 190

Ile Ala Phe Lys Met Ile Arg Thr Asn Val Ser His Val Val Gly Gln
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Leu Asp Asp Ile Arg Lys Asn Pro Arg Lys Phe Val Cys Leu Asn Asp 210 215 220

Asn Ile Asp His Asn His Lys Asp Ala Arg Thr Val Lys Ala Val Leu 225 230 235 240

Arg Asp Phe Tyr Glu Ser Met Phe Pro Ile Pro Ser Gln Phe Glu Leu 245 250 255

Pro Arg Glu Tyr Arg Asn Arg Phe Leu His Met His Glu Leu Gln Glu 260 265 270

Trp Arg Ala Tyr Arg Asp Lys Leu Lys Phe Trp Thr His Cys Val Leu 275 280 285

Ala Thr Leu Ile Ile Phe Thr Ile Phe Ser Phe Phe Ala Glu Gln Ile 290 295 300

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Ala Ser Pro Asp Arg Ile Arg Val 325

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Leu Cln Pro Lys Arg Glu Pro Ser Ala Val Ser Gly Pro Leu His Leu 50 55 60

Phe Arg Leu Ala Gly Lys Cys Phe Ser Leu Val Glu Ser Thr Tyr Lys 70 75 80

Tyr Glu Phe Cys Pro Phe His Asn Val Thr Gln His Glu Gln Thr Phe 85 90 95

Arg rp Asn Ala Tyr Ser Gly Ile Leu Gly Ile Trp His Glu Trp Glu
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Ile  $\rightleftharpoons$  le Asn Asn Thr Phe Lys Gly Met Trp Met Thr Asp Gly Asp Ser 115 120 125

Cys His Ser Arg Ser Arg Gln Ser Lys Val Glu Leu Thr Cys Gly Lys 130 135 140

Ile Asn Arg Leu Ala His Val Ser Glu Pro Ser Thr Cys Val Tyr Ala 145 150 155 160

Leu Thr Phe Glu Thr Pro Leu Val Cys His Pro His Ser Leu Leu Val 165 170 175

Tyr Pro Thr Leu Ser Glu Ala Leu Gln Gln Arg Leu Asp Gln Val Glu 180 185 190

Gln Asp Leu Ala Asp Glu Leu Ile Thr Pro Gln Gly Tyr Glu Lys Leu 195 200 205 Leu Arg Val Leu Phe Glu Asp Ala Gly Tyr Leu Lys Val Pro Gly Glu Thr His Pro Thr Gln Leu Ala Gly Gly Ser Lys Gly Leu Gly Leu Glu 235 Thr Leu Asp Asn Cys Arg Lys Ala His Ala Glu Leu Ser Gln Glu Val 245 250 Gln Arg Leu Thr Ser Leu Leu Gln Gln His Gly Ile Pro His Thr Gln 265 260 Pro Thr Glu Thr Thr His Ser Gln His Leu Gly Gln Gln Leu Pro Ile 280 Gly Ala Ile Ala Ala Glu His Leu Arg Ser Asp Pro Gly Leu Arg Gly Asn 🗓e Leu 305 📺 Д <2102 10 <211> 2070 <212> DNA <213> Mus musculus <220> <221> misc feature <222> (186) ... (186) <223> n is a, t, g, or c <4000> 10 60 qtqagaccct aggagcaatg gccgggcggc tggctggctt cctgatgttg ctggggctcg cgtcgcaggg gcccgcgccg gcatgtgccg ggaagatgaa ggtggtggag gagcctaaca 120 cattenagt agogateac ggteetgegg ettggggace gageetgget ggttettetg 180 acctantcaa ttccataggc tgaataaccc gttcttgccc caggcaagcc gccttcagcc 240 300 caagagagag ccttcagctg tatcccgcaa attaagagaa attaatttca aacgatttag aaagtattet agecaggega tgatggegea egeetttaat eecageaett gggaggeaga 360 ggcaggcaga tttccgagtt caaggccatc agaactgact gtacatctta gtacagttta 420 gcatgtgatc agagatctga atcacaaagc tgggcctgcg tggtaaagca ggtcctttct 480 aataaggttg cagtttagat tttctttctt aactctttta ttctttgaga cagggtttct 540 600 caacaqtqqq tqtcctqqaa ctcacttttq taaaccaqqc tqcccttaaa ctcacaaagc 660 totgtoagoo totgootoot gagtgotggg attaaaggto cacaccotgt toattoattt 720 ttaatttttg agactgggtc tcattatgtg gccctagaca gatactgaga gcctcctcca 780 caggaacaag catgggaatc ctgccacaga caaccagttc tqtggtctgg agatgagttt 840 gtcagtccct aggagttagg tcagcctgcc tctgcattcc caataattta ggaaaggagc

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Asn Lys Gln Thr Lys Lys Asn Met Ser Ile Ser Gly Lys Glu Leu Ala 50 55 60

Ile Ser Pro Ala Tyr Leu Leu Trp Asp Leu Ser Ala Ile Ser Gln Ser 65 70 75 80

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Leu

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Ala Thr Trp Ser Asp Arg Glu Val Arg Thr Phe Leu Thr Arg Ile Tyr

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Leu Ala Ala A 435	Asn Phe	Ala Val	Arg Pro	Lys Tyr	Asn Phe 445	His Val	Ser
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Thr Glu Val V	Val Glu	Ser Leu 470	Asp Arg	Leu Arg 475	Arg Asn	Pro Arg	Lys 480
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Gln Gly Leu Ala Phe Leu Ser Gly Phe Pro Pro Thr Phe Lys Glu Thr

Ser Gln Leu Lys Thr Lys Leu Pro Arg Lys Ala Phe Pro Leu Lys Ile 245

Lys Leu Leu Arg Leu Tyr Ser Glu Ala Ser Val Ala Leu Leu Lys Leu 265

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